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Case Report

Secondary Aneurysmal Bone Cyst Simulating Malignant Transformation in Fibrous Dysplasia

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Fibrous dysplasia is a benign bony lesion that may simulate a bone malignancy.[1] Rarely, fibrous dysplasia can have a superimposed aneurysmal bone cyst and an aggressive appearance on radiographs. This may suggest sarcoma arising in fibrous dysplasia.[2]

This article reports a long-standing fibrous dysplasia in which a lytic component developed.

CASE REPORT

In 1965, a 41-year-old woman presented with pain in her thigh. Medical history was unremarkable. Laboratory tests were normal. Radiographs of the proximal femur showed a well-marginated lytic lesion. Radiographic diagnosis was fibrous dysplasia (Figure 1). The patient was not treated; however, she underwent follow-up.

Fifteen years later, radiographs showed a well-marginated "ground-glass" radiodensity; the size was stable (Figure 2). In July 1995, the patient reported pain in the proximal thigh. Standard radiographs revealed a lytic lesion in the proximal left femur, with indistinct borders thinning the cortex (Figure 3). In October, the patient had a left subtrochanteric pathologic fracture, initially treated at another institute with a plaster cast (Figure 4).

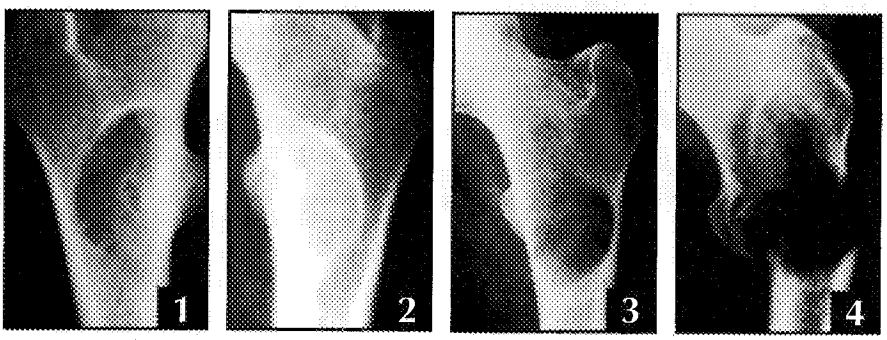

Figure 1: Radiograph taken in 1965 shows a lytic intertrochanteric area with well-defined borders. **Figure 2:** Fifteen years later, there was increased calcifying with prominent "ground-glass pattern" well-defined margins. **Figure 3:** Thirty years later, a lytic extensive lesion appeared in the same area. Thinning of the cortex along with indistinct borders was detected. **Figure 4:** After pathologic fracture, the nature of the lesion was im-possible to assess. The differential diagnosis was between sarcoma in fibrous dysplasia and aneurysmal bone cyst in fibrous dysplasia.

A needle biopsy and two incisional biopsies were obtained and the histologic diagnosis was fibrous tissue and blood. After frozen section, the diagnosis of fibrous dysplasia with cystic aspect was made. At the time of surgery, a cystic area was identified. It was externally covered by fibrous tissue, most likely fracture callus. The inner part of the cyst showed tissue that had a layer of brown color and rubbery consistency. The patient underwent osteosynthesis surgery with a titanium plate device and received autoplastic grafts taken from the left wing of the iliac crest (Figure 5).

On histologic examination, there was spindle cell proliferation in a collagenous matrix. The spindle cells (fibroblasts) had no cytologic atypia. Woven bone trabeculae without osteoblastic rimming were scattered throughout (Figure 6). Occasionally, small collections of foam cells were identified. Engrafted in this tissue was a cyst containing blood with a granulation tissue wall and extensive fibrosis. Scattered giant cells were embedded in the fibrous granulation tissue of the cyst wall (Figure 7).

Histopathologic diagnosis was fibrous dysplasia with secondary aneurysmal bone cyst. The differential diagnosis according to radiographs included secondary malignancy in fibrous dysplasia and fibrous dysplasia with

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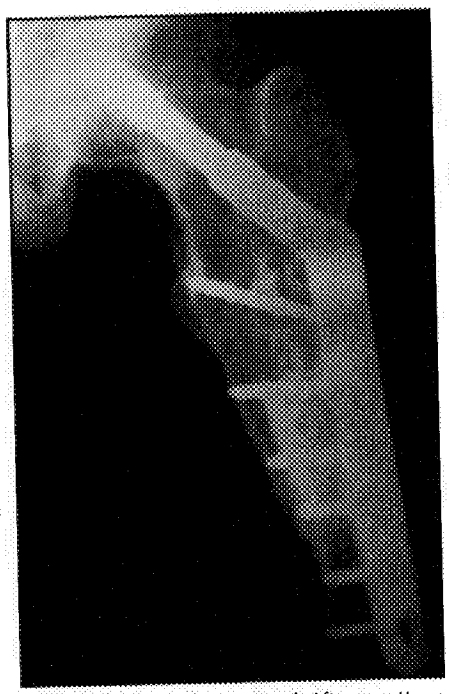

Figure 5: Postoperative control. After curettage and bone graft, a plate device was implanted.

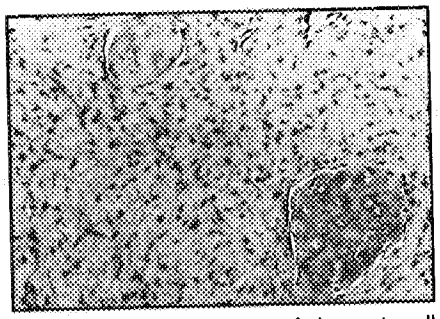

Figure 6: Low-power view of the cyst wall spindle cells arranged in short fascicles with scattered small vessels. Fibrin and blood fill up the cystic space (hematoxlyin-eosin; original magnification ×6).

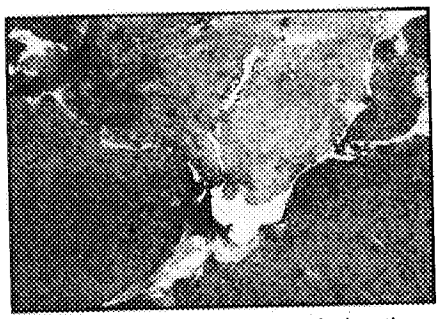

Figure 7: High-power view of the tissue adjuvant to the cyst. Plump spindle cells with benign cytologic appearance surrounding small woven bony trabeculae without osteoblastic rim (hematoxlyin-eosin; original magnification ×300).

secondary aneurysmal bone cyst and pathologic fracture with hematoma.

The patient's postoperative course was unremarkable, and 24 months postoperatively, the patient remains well without evidence of disease (Figure 8).

DISCUSSION

Fibrous dysplasia is hamartomatous malformation[3] characterized by an intramedullary cavity proliferation of fibrous tissue[1-3] with woven bony trabeculae. Fibrous dysplasia can be monostotic or polyostotic. The polyostotic form is less frequent than monostotic fibrous dysplasia. The most common locations of polyostotic fibrous dysplasia are the lower extremities and pelvis. Polyostotic form may be associated with Albright's syndrome (cutaneous pigmentation, endocrine disorders, precocious puberty in females, premature skeletal maturity, and bone deformity).[2-7] The monostotic form is more frequent, affecting individuals aged 5-20 years.[2,3] It can be asymptomatic or painful and swelling may be present.

Radiographic features are related to the woven bone production and its mineralization. The lesion is a well-marginated lytic intraosseous defect with a ground-glass appearance.[3,8]

Our patient had the radiographic appearance of fibrous dysplasia at the time of symptom onset. The radiographic aspect was consistent with fibrous dysplasia in the following 30 years.

The sudden, rapid disappearance of bone structures and cortical destruction after pathologic fracture first suggests malignant change in fibrous dysplasia. This change occurs in 4%-10% of cases of fibrous dysplasia, and histologically, the malignancy has the features of osteosarcoma, fibrosarcoma, or malignant fibrous histiocytoma.[9-11] Furthermore, benign lesions such as aneurysmal bone cyst can be secondary in fibrous dysplasia and can be aggressive and may simulate a sarcoma.[12-17]

In our patient, the preoperative diagnosis of possible malignant transformation of fibrous dysplasia prompted biopsy. After three negative biopsy procedures, on frozen section the diagnosis was fibrous dysplasia with associated aneurysmal bone cyst. Imaging studies such as computed tomography (CT) or magnetic resonance imaging were unable to differentiate sarcomatous transformation of fibrous dysplasia from secondary aneurysmal bone cyst in fibrous dysplasia. However, plain radiographs and CT were helpful in assessing the evolution of the lesion.[18-20]

Histologic study of the lesion was important for the diagnosis and helped avoid radical surgery. Fibrous dysplasia associated with aneurysmal bone cyst is not a rare condition.[14,16] In 1986, Diercks et al[12] reported a case of aneurysmal bone cyst in association with polyostotic fibrous dysplasia. They pointed out that nine similar cases were reported in the literature. Four cases were in the jaw, one in a rib, one in the tibia, and three were unspecified. The case reported by Diercks et al[12] was considered the first report of aneurysmal bone cyst in fibrous dysplasia in the femur.

Fibrous dysplasia can cause cortical thinning and bony expansion but rarely does it focally destroy and extend through cortical bone. If this is detected occasionally in fibrous dysplasia involving the flat bones and narrow bones (ribs), it is rare in the long bones.[21] In this clinical setting, a secondary malignancy or secondary aneurysmal bone cyst has to be considered in the differential diagnosis.[22-25]

Rarely, a fibrous dysplasia-like lesion in the metaphysis of long bone grows aggressively causing bony expansion, cortical destruction, and extension toward the epiphysis. In this case, the histology may be bland with hypocellular spindle cell areas and low mitotic rate. The diagnosis of low-grade central osteosarcoma has to be considered.[26]

The case presented deals with a lytic lesion in the proximal femur with frac-

ture in a long-standing fibrous dysplasia. It was considered worrisome for malignancy. The biopsy clarified the benign nature of the lesion. This case suggests that aneurysmal bone cyst engraftment on long-standing monostotic fibrous dysplasia may simulate a malignancy. This possibility has to be considered when aggressive features are associated with a benign lesion. The diagnosis of benign lesion will avoid overtreatment of the patient.

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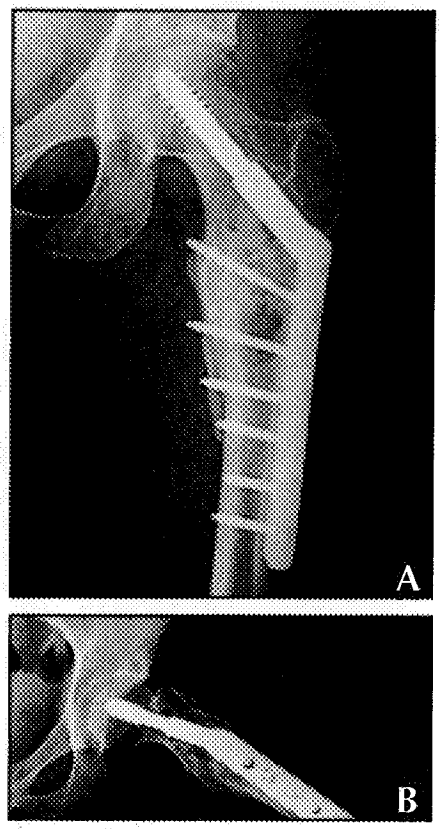

Figure 8: Radiographs show no evidence of disease, autograft fusion (A), and plate stability (B).